May-Thurner syndrome: a not so uncommon cause of a common condition

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May-Thurner syndrome is a rarely diagnosed condition in which patients develop iliofemoral deep venous thrombosis (DVT) due to an anatomical variant in which the right common iliac artery overlies and compresses the left common iliac vein against the lumbar spine. This variant has been shown to be present in over 20% of the population; however, it is rarely considered in the differential diagnosis of DVT, particularly in patients with other risk factors. Systemic anticoagulation alone is insufficient treatment, and a more aggressive approach is necessary to prevent recurrent DVT. Herein, we present a patient with multiple risk factors for DVT. With a comprehensive diagnostic approach, she was found to have May-Thurner syndrome. Local infusion of thrombolytics as well as mechanical thrombectomy failed to resolve the thrombus. Subsequently the patient underwent successful stent placement in the area that was compressed followed by 6 months of chronic anticoagulation with warfarin. There has been no recurrence of DVT in the ensuing 18 months.

27-year-old African American woman presented with a 12-hour history of left lower extremity swelling associated with 7/10 dull throbbing pain. The patient stated that the pain began initially in her left foot and, within several hours, began to radiate to her left groin. These symptoms began 2 weeks after an 8-hour automobile ride from Houston to New Orleans. The patient had no prior medical history. Her only daily medication was an oral contraceptive, drospirenone/ ethinyl estradiol. She had no significant family history. She smoked several cigarettes per month but denied any alcohol or drug use. On examination, the patient's vital signs were within normal limits. Her left lower extremity was warm, swollen, and erythematous from mid-calf to mid-thigh. Dorsalis pedis and posterior tibial pulses were normal, and reflexes, strength, and sensation were all normal. A complete blood count and basic metabolic profile were within normal limits. Her prothrombin time was 16.5 seconds, activated prothromboplastin time was 26.3 seconds, and international normalized ratio was 1.3. The patient had an elevated D-dimer at 24.3 mg/L.

Left lower extremity ultrasound revealed extensive iliofemoral deep venous thrombosis (DVT). Urgent lower extremity venography revealed extensive thrombus in the left femoral, popliteal, and iliac veins (*Figure 1*). A 5 French Cragg-McNamara catheter was inserted in the left femoral vein,

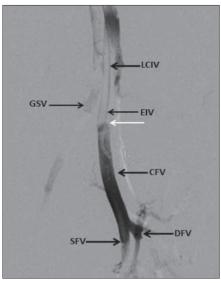


Figure 1. Thrombus appearing as a filling defect in the iliofemoral vein (proximal to the white arrow). The site of dye injection was distal to SFV and DFV bifurcation. LCIV indicates left common iliac vein; EIV, external iliac vein; GSV, greater saphenous vein; CFV, common femoral vein; DFV, deep femoral vein; SFV, superficial vein.

through which an 18-hour infusion of 1 mg/hr tissue plasminogen activator (tPA) was initiated. Following the tPA infusion, the patient was started on a heparin drip, which was titrated to an activated prothromboplastin time of 50 to 70 seconds. Repeat venography 2 days later revealed residual thrombus and stenosis (*Figure 2*). A mechanical thrombectomy was performed using a Trellis device (*Figure 3*) with the simultaneous infusion of an additional 5 mg of tPA, after which some resolution of the thrombus was noted; however, stenosis was still present (*Figure 4*). The stenosis within the left common iliac vein was then dilated with a 10×40 mm balloon, and a 14×40 mm stent was placed across the stenotic area (*Figure 5*). While the right common iliac artery was not directly visualized, this patient was proven to have May-Thurner syndrome (MTS) based on the proximal location of her DVT, the stenosis in the left common iliac vein

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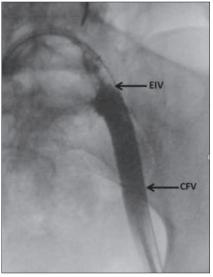


Figure 2. Residual left iliofemoral thrombus and stenosis (proximal to the EIV arrow) noted after tissue plasminogen activator and heparin administration. EIV indicates external iliac vein; CFV, common femoral vein.



Figure 3. Trellis device (arrows) used for mechanical thrombectomy.

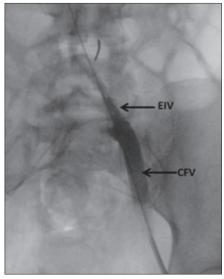


Figure 4. Left iliofemoral thrombus (proximal to the EIV arrow) improved after mechanical thrombectomy but with residual stenosis. EIV indicates external iliac vein; CFV, common femoral vein.

in an area directly overlying the lumbar spine, and the appearance of residual thrombus and stenosis following thrombolytic administration and thrombectomy.

Following placement of the stent, the patient was started on 5 mg of daily warfarin, for which she was bridged with twicea-day injections of 1 mg/kg enoxaparin for 5 days. The patient discontinued the warfarin after 6 months when a hypercoagulability workup, consisting of tests for Factor V Leiden, prothrombin gene mutation, homocysteine level, and antithrombin III level, was negative. Eighteen months postoperatively, the patient has had no recurrence of DVT.

DISCUSSION

MTS was first described in 1957 when it was noted that 22% of 430 cadavers on autopsy possessed an anatomical variant in which an overriding right common iliac artery caused compression of the left common iliac vein against the lumbar spine (1). More recently, a similar prevalence (22%–24%) of MTS was reported in a retrospective analysis of computed tomography scans (2). This compression is associated with intimal hyperplasia, which creates the potential for venous stasis and subsequent thrombosis (1). Despite the relatively high incidence of this anatomical variation, the clinical prevalence of MTS-related DVT is surprisingly low, reportedly occurring in only 2% to 3% of all lower extremity DVTs (3). It is thought that this low occurrence rate may be an underestimate of the actual prevalence due to missed diagnoses; the fact that there is a 55.9% predominance for left-sided DVT would seem to support this notion (4).

One reason for the apparent underdiagnosis of MTS may be the prevalence of other more easily recognized risk factors for DVT. DVT is more common in women, and 72% of women diagnosed with MTS are relatively young (aged 25-50) (3, 5). Additionally, these patients often have a history of oral contraceptive

use, recent pregnancy, or recent prolonged travel. Accordingly, in a patient with identifiable risk factors, the diagnostic workup is often halted once the diagnosis of DVT is confirmed. Failure to correct the anatomic substrate of MTS could lead to DVT recurrence and additional complications,

including pulmonary emboli, chronic venous stasis, and iliac vein rupture (28%

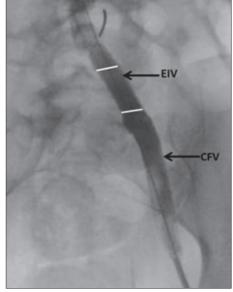


Figure 5. Installation of smart stent (between white bars) shows appearance of dye proximal to the previous area of stenosis. EIV indicates external iliac vein; CFV, common femoral vein.

of patients with iliac vein rupture have MTS) (6, 7).

The anatomical defect associated with MTS occurs high in the pelvis, an area that is not easily visualized by ultrasound (8). Accordingly, if MTS is suspected, contrast venography, magnetic resonance imaging, or intravascular ultrasound should be performed (9). Following thrombus removal, a computed tomographic angiogram or magnetic resonance venography should be obtained to assess the degree of stenosis and the hemodynamic effects of iliac vein compression (9).

It is generally accepted that long-term anticoagulation, while indicated, is not adequate to prevent long-term sequelae in MTS patients and that a more invasive therapeutic approach is indicated (5). Several historical innovative techniques have included creation of tissue slings, retropositioning of the overriding vessel, and venovenous bypass (10–12). The mainstay of therapy has traditionally involved open repair of the affected vein; however, the standard of care has since evolved into a hybrid approach, involving the combination of thrombolytics and endovascular intervention. Both Moudgill et al and Suwanabol et al recommend catheter-directed thrombolysis combined with percutaneous mechanical thrombectomy (5, 9). It has also been suggested that an inferior vena cava filter be placed prior to lower extremity intervention in order to prevent further embolization during lytic therapy, especially in individuals with large clot burdens (5).

It is typically recommended that, following initial clot lysis, thrombolytic infusion should be continued for an additional 24 to 48 hours (9). Following completion of thrombolytics, an intravascular stent should be deployed in the area of iliac vein compression. Repeat imaging should be obtained to verify that the stent is positioned across the entire area of the compressed vein. Suwanabol et al recommend the use of large (12-14 mm) self-expanding stents, placed across the extent of the stenosis and extending into the inferior vena cava, if possible, to prevent migration (9). Stent placement has proven highly successful in MTS, with 2-year iliac vein patency rates reported between 95% and 100% (13). Following stent placement, systemic longterm anticoagulation is recommended for at least 6 months (5). In our patient, we decided to discontinue warfarin therapy after 6 months for two reasons. First, we believed that the risks of chronic anticoagulation outweighed the benefits, since the underlying anatomical defect had been corrected by a stent. Second, the discontinuation of warfarin allowed us to test for hypercoagulability, which may have increased the risk for future DVT.

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